Review Article

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Oral Manifestations of Sjogren's Syndrome

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Introduction

Sjogren's Syndrome is an autoimmune rheumatic disease characterized by focal mononuclear cell infiltration of the salivary and lacrimal glands.[1] It is characterized by oral and ocular dryness, exocrine dysfunction, lymphocytic infiltration, and destruction of exocrine glands.[2]

Sjogren's Syndrome is subclassified into Primary and Secondary, in which Primary Syndrome consists of dryness of the mouth (xerostomia) and eyes (keratoconjunctivitis sicca), while Secondary Syndrome is associated with other systemic rheumatic diseases, such as rheumatoid arthritis, lupus, or scleroderma.[3].

Etiology and Pathogenesis

The exact etiology of Sjogren's syndrome is unknown; it is considered to be multifactorial. It is mainly due to continuous stimulation of the autoimmune system. The immune cells involved are B and T lymphocytes, even though the mechanisms underlying humoral and cell-mediated reactions are unknown.

1. GENETICS: The genetic predisposition to Sjogren's syndrome has been reported because of multiple reports of two or more members of the same family developing a syndrome.

2. ENVIRONMENT: Infection caused by Epstein-Bar & Helicobacter pylori bacteria is considered a possible etiological factor for initiating Sjogren's syndrome. Several viruses have been implicated as possible environmental triggers of SS, including EBV, CMV, HHV-8, etc.

3. SEX HORMONE: The majority of people affected with SS are females, compared to males, suggesting that sex hormones play a role in an autoimmune response. Oestrogen is an immune stimulator that has a role in lymphocyte growth, differentiation, proliferation, antigen presentation, cell survival, and apoptosis. It declines during menopause, and this is when women are most susceptible to developing SS, suggesting that either oestrogen decline or the difference in the oestrogen: androgen ratio is involved with disease onset.

4. INFLAMMATORY REACTIVITY: The result of an external response can also stimulate inflammation. Lymphocytes most often accumulate in the salivary glands and produce several pro-inflammatory cytokines, therefore sustaining the disease.

European-American Consensus Group Criteria for SS[4]

1. Ocular symptoms (positive response to atleast one of three)
   - Daily, persistent, troublesome eyes for more than three months
   - The recurrent sensation of sand or gravel in the eyes.
   - Use of tear substitutes more than three times per day.
2. Oral symptoms (positive response to atleast one of three)
   - A daily feeling of dry mouth for more than three months.
   - Recurrent or persistently swollen salivary glands as an adult.
   - Frequent drinking of liquids to aid in swallowing food.
3. Ocular signs (Positive result for atleast one of 2 tests)

- Schirmer's test, performed without anesthesia (≤ 5mm in 5 min)
- Rose Bengal score or other ocular dye scores (≤ 4 according to van Bijsterveld's scoring system)

4. Histopathology in Minor salivary gland biopsy
   - Focal lymphocytic
   - sialoadenitis, with focus score≥1 (a focus is defined as ≥50) lymphocytes per 4mm^2 of glandular tissue adjacent to normal-appearing mucous acini)
5. Salivary gland involvement

   (the positive result of one and few)
   - Unstimulated whole salivary flow ≤ 1.5 ml/ 15 min)
   - Parotid sialography shows the presence of diffuse sialectasis (punctuate, cavitary or destructive pattern) without evidence of obstruction in the major ducts.
   - Salivary scintigraphy shows delayed uptake, reduced concentration, and / or delayed excretion of tracer.
6. Autoantibodies.
   a. Presence in the serum of antibodies to Ro (SS-A) or La (SS-B) antigens, or both.

Classification Criteria

- Primary Sjogren's Syndrome

  The presence of any four-six items as long as either item 4 (Histopathology) or item 6 (Serology) is positive

  The presence of any three of the four objective criteria(items 3,4,5 and 6)
- Secondary Sjogren's Syndrome

  In the presence of another connective tissue disease, the presence of item 1 or item 2, plus any two from items 3, 4 and 5

Exclusion Criteria

Past head and neck radiation treatment.

Hepatitis infection.

AIDS.

Pre-existing lymphoma.

Sarcoidosis.

Use of anticholinergic drugs (Since a time shorter than fourfold the half-life of the drug).

Oral Manifestations

- Patients with SS experience the full spectrum of oral complications that result from the decreased salivary function.[5]
- All patients complain of dry mouth and attendant difficulties in speaking,tasting, and swallowing, and the need to sip liquids throughout the day.
- **Xerostomia** may become evident with nocturnal awakening with thirst and the need to have chewing gum or lozenges to stimulate saliva production.
- **Dental caries** are the most common clinical manifestation. Specifically, **root and incisal caries**, seldom seen amongst the general population, are a greater concern for Sjogren's syndrome. It does not reside with periodontitis.
- Other subjective complaints include dysgeusia or hypogeusia, coughing episodes, and choking.
- The mucosa may be painful and sensitive to spices and heat. Patients often have dry, cracked lips and angular cheilitis. Intraorally, the mucosa is pale and dry, friable, or furrowed; minimal salivary pooling is noted, and the saliva that is present tends to be thick and ropy.
- The tongue is often smooth (depapillated) and painful.
- **Mucocutaneous candidal infections** are common, particularly in the **erythematous** form.
- An oral burning sensation, stomatitis, or glossodynia is a common complaint in SS patients. It is secondary to a fungal infection.

- Due to the lack of lubricating saliva, traumatic or frictional injury is increased.
- Removable prosthesis are less well tolerated due to the reduction in retention usually afforded by saliva.
- Bilateral Parotid gland enlargement has been found in 25%-60% of all patients.[6]

Treatment

- Since Sjogren's syndrome has no cure, treatment is symptomatic and supportive. Without sufficient saliva to restore the oral pH and regulate microbial populations, the mouth can rapidly colonize deleterious bacterial, viral, and fungal populations. A personalized treatment plan must be developed for the patient by their health care professionals to treat the various symptoms. A preventive oral health plan should include meticulous oral hygiene instructions to improve quality of life and avoid complications.
- Mechanical tooth brushing 2 to 3 times daily with a prescription fluoride gel containing 1.1% sodium fluoride or remineralizing dentifrice. Interdental aids such as a Waterpik or proxy-brush in addition to flossing
- Dietary counseling
- A complement of chemotherapeutic agents
- More frequent recall care, 3 to 4month.
- Water is the most easily available and commonly used home remedy for managing discomfort related to xerostomia by producing some moisture to the oral mucosa and often helping in speech and swallowing.[7,8]
- Professional application of topical 5% sodium fluoride varnish and daily home fluorides such as 1.1% sodium fluoride prescription dentifrices are preventive strategies that decrease microbe colonization and strengthen tooth enamel—thus making tooth surfaces more resistant to caries
- Oxygenated Glycerol Triester saliva substitute spray was found to be more effective than water-based electrolyte spray.
- Since no commercial saliva substitute has been developed which accurately replicates all essential qualities of natural saliva, attempts should be made to increase the natural flow of saliva as much as possible. Sugar-free gum, mints, and lozenges are advisable in those with the residual capacity to encourage increased salivary production. [9]
- Sjogren's syndrome is associated with the increased occurrence of candida infections. Treatment includes topical antifungal agents and may be followed by systemic antifungal agents for persistent or recurrent episodes. A typical regimen includes antifungal cream and a pastille, troche, or oral suspension of an antifungal 3 to 5 times daily for one week, followed by systemic treatment with an azole.[10]
- To prevent candidiasis, wearing dentures overnight should be discouraged; dentures should be cleaned and treated daily with benzoic acid, 0.12% Chlorhexidine Gluconate or 1% sodium hypochlorite, as primary Sjögren's syndrome carries a high risk of oral candidiasis and a high frequency of multiple candida infections.[11]

Conclusion

As Sjogren's syndrome affects many organs, the treatment for this involves a multidisciplinary approach. It should be diagnosed earlier and treated promptly.

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